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Title **The Clinical Effectiveness and Safety of Prophylactic Retinal Interventions to Reduce the Risk of Retinal Detachment and Subsequent Vision Loss in Adults and Children With Stickler Syndrome: A Systematic Review**

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Aim

To assess the evidence for the clinical effectiveness and safety of primary prophylactic interventions for preventing retinal detachment (RD) in previously untreated eyes without RD in patients with Stickler syndrome.

Conclusions and results

The literature search identified 1444 unique citations, of which 4 studies satisfied the inclusion criteria. The 2 principal studies were retrospective cohort studies with control groups in populations with type 1 Stickler syndrome. One study evaluated 360° cryotherapy (n=204) and the other focal or circumferential laser treatment (n=22). Both studies reported a statistically significant difference in the rate of RD per eye between the groups receiving prophylaxis and the controls. However, both studies were subject to a high risk of bias. The results of the two supporting studies of Wagner-Stickler patients were either relatively inconsistent or unreliable. No study reported any major or long-term complications associated with the interventions. Despite the weaknesses of the evidence, the rate of RD in the intervention groups, especially the cryotherapy group, was lower than the rate either experienced in the study control groups or reported in other studies of untreated Stickler syndrome populations not exposed to prophylaxis. Only 360° cryotherapy and focal and circumferential laser treatment have been evaluated for the type 1 Stickler syndrome population, and then only by a single retrospective, controlled, cohort study in each case. Both of these studies report a significant difference between intervention and control groups (principally no treatment) and no major or long-term side effects or complications. Since both studies have a high risk of bias, the relative effectiveness of either intervention is uncertain.

Recommendations

See Executive Summary link www.hta.ac.uk/project/2156.asp.

Methods

We systematically reviewed the evidence for the clinical effectiveness and safety of primary prophylactic interventions in preventing RD in previously untreated eyes without RD in patients with Stickler syndrome. The primary outcome of interest was RD postprophylaxis. An information specialist searched 11 databases for published and unpublished literature. No restrictions were placed on language, date, or study design (other than requiring that studies have a comparator group). Two reviewers double-screened all titles and abstracts of the citations retrieved by the search to identify studies that satisfied the inclusion criteria. Any disagreements were resolved by discussion or reference to the full paper. Both reviewers independently extracted and quality assessed all included studies. The references of these studies were checked for further relevant citations. The authors of any studies with potential but unspecified Stickler syndrome patients in their study sample were contacted to retrieve any further data on the efficacy of interventions in this population.

Further research/reviews required

A service priority is to determine reliably the prevalence of Stickler syndrome, ie, how many individuals have type 1 or type 2 Stickler syndrome, and their risk of retinal detachment and subsequent blindness. A nonrandomized, prospective, cohort comparison study, in which eligible participants are treated, followed-up, and analyzed in one of three study arms (no treatment, laser therapy, or cryotherapy) would potentially enhance certainty about the relative efficacy of prophylaxis versus no prophylaxis and cryotherapy versus laser therapy. Alternatively, continued follow-up and analysis of existing study data, and data collection from relevant sample populations, are required to assess the long-term risks of blindness, retinal detachment, and prophylaxis.

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